

CASES FOR DIAGNOSIS

Hyperkeratotic Papules on the Hands

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Clinical History

The patient was a 24-year-old black woman with no past history of interest, who was seen for asymptomatic lesions on the margins of both hands and in the creases of both palms. The lesions had progressively increased in number over a period of 2 years. In her family history, it was found that her father and 1 brother had similar lesions on the margins of the hands and feet.

Physical Examination

Physical examination revealed crateriform, polygonal, hyperkeratotic papules on the margins of both hands (Figure 1A), in the creases of both palms (Figure 1B), and on the palmar aspect of the interphalangeal joints (Figure 2A), where callous-type lesions were also present (Figure 2B). No other areas of the body were affected.

Histopathology

One of the papular lesions was biopsied and histopathological study revealed an area of orthokeratotic hyperkeratosis on a depressed region of epidermis that was slightly acanthotic but otherwise normal. The underlying dermis was considered normal (Figure 3).

What Was the Diagnosis?

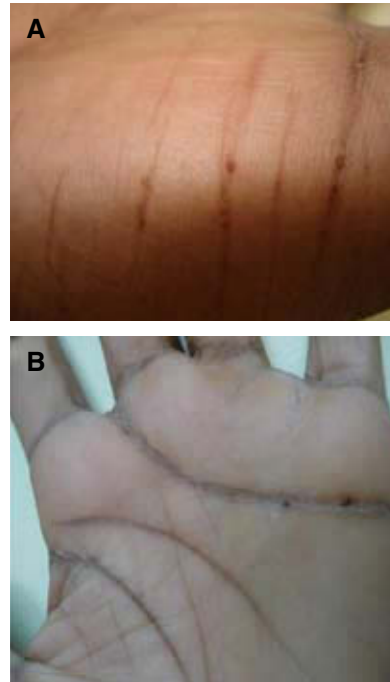


Figure 1.

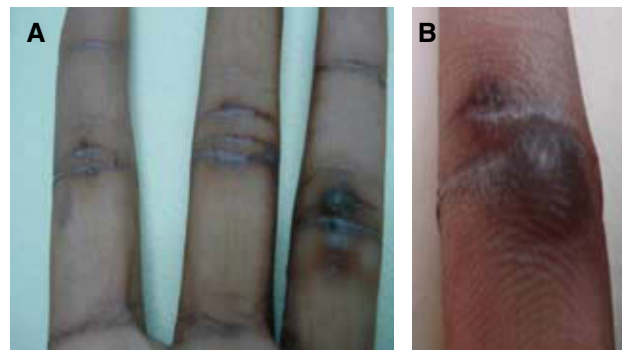


Figure 2.

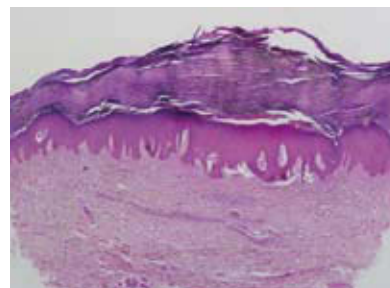


Figure 3.
Hematoxylin-eosin,
×40.

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Diagnosis

Focal acral hyperkeratosis

Discussion

The term focal acral hyperkeratosis was proposed by Dowd and collaborators in 1983. They reported 15 patients who presented small, oval or polygonal, hyperkeratotic papules on the medial and lateral margins of the hands and feet. All but one of the patients were black and 12 were women. There was a family history of similar lesions in approximately half of the patients. Histopathological study revealed an area of orthokeratotic hyperkeratosis on a crateriform depression of the epidermidis, which was slightly acanthotic. There were no inflammatory cells in the dermis and the collagen and elastic fibers were of normal appearance.¹ Dowd and collaborators² proposed the term focal acral hyperkeratosis to distinguish this condition from classical acrokeratoelastoidosis, which has similar clinical and histopathological findings but, in contrast to focal acral hyperkeratosis, presents dermal elastorrhexis.

Focal acral hyperkeratosis is a rare disorder that occurs most commonly in black women. Onset typically occurs during the second or third decade of life, and the number of lesions gradually increases over the years.³

Focal acral hyperkeratosis and acrokeratoelastoidosis usually present sporadically, though there are cases with a family history suggestive of an autosomal dominant inheritance with incomplete penetration. The etiology is unknown, though a primary disorder of elastic fibers has been proposed. The genetic mutation has been linked to chromosome 2. There is no reliable evidence that exposure to sunlight, trauma, or arsenic ingestion are involved in its pathogenesis and controversy still exists over whether focal acral hyperkeratosis is an independent disorder or forms part of the spectrum of acrokeratoelastoidosis. Among

family members, changes in the elastic fibers found in one patient may not be present in the others. However, ultrastructural studies have demonstrated alterations of the elastic fibers in some cases of focal acral hyperkeratosis, suggesting that cases with subtle changes would not be detected in the histopathological examination.⁴

Other dermatological disorders that can be confused with focal acral hyperkeratosis include degenerative collagenous plaques of the hands, flat warts, acrokeratosis verruciformis, porokeratosis punctata palmaris et plantaris, palmoplantar lichen planus, and connective tissue nevus.⁵

Diagnosis is based on the history and the typical clinical pattern, the absence of abnormalities of the collagen and elastic fibers on histological examination, and a predominance in black women.¹

Treatment is generally unnecessary as the lesions are asymptomatic. Numerous options have been tried, such as liquid nitrogen, salicylic acid, and methotrexate, but without success. Only etretinate has shown some efficacy, although recurrence occurs on withdrawal of the treatment.^{3,5}

Conflicts of interest

The authors declare no conflicts of interest.

References

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